Impact of Re-Coarctation Following the Norwood Operation on Survival in the Balloon Angioplasty Era

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OBJECTIVES The objective of this study was to determine the efficacy of balloon angioplasty (BA) by comparing the immediate and long-term outcomes of patients with and without re-coarctation after a Norwood procedure.

BACKGROUND Although BA has become the standard means for treating recurrent coarctation following a Norwood operation, it has been suggested that re-coarctation remains a significant cause of morbidity and mortality.

METHODS Patients who survived a Norwood operation from December 1986 through June 2001 were studied. Differences between groups were evaluated by t test and logistic regression. Survival differences were tested by log-rank tests using Kaplan-Meier survival curves.

RESULTS Fifty-eight of 633 patients underwent treatment for re-coarctation (9.2%). Thirty-five patients underwent BA (before 1988, 23 had surgery). Median age at catheterization was 6.6 months (1.9 to 35.6 months). Balloon angioplasty was successful (gradient <10 mm Hg) in 32 of 35 patients (92%). There were no BA-related deaths or neurologic complications. Recurrent obstruction after BA occurred in seven patients (20%); five underwent re-dilation. Kaplan-Meier estimates of freedom from recurrent obstruction after initial BA were 97% at one month, 79% at one year, and 79% at five years. There were no differences in survival between patients with re-coarctation treated by BA and patients who did not undergo treatment for re-coarctation.

CONCLUSIONS We found that 9.2% of patients underwent treatment for re-coarctation following a Norwood operation. Balloon angioplasty is effective, with low morbidity, no early mortality, and no difference in long-term survival when compared with patients who did not have re-coarctation. Recurrent coarctation following BA occurred in 17% of patients, usually within the first year after BA. (J Am Coll Cardiol 2005;45:1844–8) © 2005 by the American College of Cardiology Foundation

Re-coarctation is common in patients who have undergone a Norwood operation for hypoplastic left heart syndrome (HLHS), with a reported incidence from 11% to 37% (1–7). After the efficacy of balloon angioplasty (BA) was demonstrated for native and postoperative aortic arch obstruction, the procedure was applied to patients with HLHS (8–12). Subsequently, transcatheter BA has become the standard means for treating recurrent coarctation in this setting. Several series have defined the short-term efficacy of this treatment; however, recurrent obstruction following balloon dilation is not infrequent, and information regarding the long-term outcome of these patients has not yet been addressed.

The purpose of this study is to examine the impact of BA on morbidity and mortality when used as the primary modality of treatment for re-coarctation following the Norwood operation. First, we evaluated all patients who underwent angioplasty for re-coarctation after a Norwood operation to identify the incidence of and risk factors for development of re-coarctation of the neo-aorta in patients after the Norwood operation. Second, we investigated the efficacy of BA for treatment of re-coarctation and evaluated the risk of recurrent obstruction. In doing so, we examined potential markers to predict initial success and risk factors for recurrent obstruction as well as the efficacy of repeat BA. Last, we evaluated the impact of re-coarctation by comparing long-term survival in patients with re-coarctation treated by BA with survival of patients who underwent a Norwood procedure for HLHS but did not develop subsequent arch obstruction.

METHODS

Patient population. We reviewed the surgical database at The Children’s Hospital of Philadelphia to identify all patients who underwent a Norwood operation for HLHS from December 1986 through June 2001 and survived the initial postoperative period (>48 h). The diagnosis of HLHS was based on echocardiographic evidence of a diminutive ascending aorta, aortic atresia or stenosis, and a
hypoplastic left ventricle. Patients undergoing a Norwood operation for double-outlet right ventricle with mitral atresia and aortic arch hypoplasia, as well as common unbalanced atroventricular canals with left ventricular hypoplasia-associated arch hypoplasia, were also included in this study. The institutional review board of the Children’s Hospital of Philadelphia approved this retrospective study.

Patients presented for cardiac catheterization based on clinical suspicion of arch obstruction or as a routine procedure in anticipation of their next surgical procedure. The catheterization laboratory database was reviewed to identify all patients with re-coarctation after a Norwood operation. In general, patients were diagnosed with re-coarctation if they had a >20 mm Hg gradient and >50% stenosis. Individuals with significant angiographic obstruction and lower gradients in the setting of poor ventricular function were also included. The decision to treat arch obstruction was a clinical one made at the time of evaluation in each case. Those who underwent a Norwood procedure and did not develop recurrent arch obstruction served as the control population in this study. Variables such as weight at the time of the Norwood operation, the presence of native aortic atresia, and the year during which the Norwood operation occurred were examined to identify possible risk factors for developing recurrent arch obstruction.

**BA technique.** All BA procedures occurred under general anesthesia, both arterial and venous accesses were secured, and heparin (100 U/kg) was administered. Standard hemodynamic data were obtained. Biplane aortography was performed using one of two projections: 1) straight anterior-posterior/lateral projections or 2) right anterior oblique with caudal angulation (Fig. 1). Balloon size was determined by the caliber of the diameter of the descending aorta at the level of the diaphragm, and balloon diameters up to 110% of this diameter were employed. Balloons of varying inflation pressures were used at the discretion of the cardiologist performing the intervention. In the majority of instances, BA was performed via an anterograde approach, although there were several cases when the arch was dilated in a retrograde manner. Post-dilation angiograms and procedural records were reviewed to identify all procedural complications.

**BA/immediate success.** Before and after procedure hemodynamic measurements were reviewed to determine the success of the intervention. We considered BA to be successful if the final gradient was ≤10 mm Hg. Multiple variables were examined to determine predictors of success and included: 1) the patient’s age and weight at the time of the BA; 2) the maximum peak systolic gradient pre-BA; 3) the minimum diameter of narrowing as well as the percent stenosis compared to the distal aorta at the level of the diaphragm; 4) the size of the balloon catheter relative to the region of narrowing; 5) the approach (anterograde vs. retrograde) used to perform the BA; and 6) the inflation pressure (high, >6 atm vs. low, ≤6 atm) of the angioplasty balloon.

**Incidence of recurrent obstruction.** Recurrent coarctation following initial BA was defined by a persistent peak systolic gradient across the arch >10 mm Hg. Factors from the initial BA procedure examined to identify possible predictive variables for recurrent obstruction included: 1) body surface area at the time of initial presentation; 2) the presence of native aortic atresia; 3) the initial transaortic gradient; 4) the initial percent stenosis; 5) the balloon inflation pressure; 6) the ratio of the diameter of the balloon catheter to narrowest region of stenosis as well as to the diameter of the aorta at the level of the diaphragm; 7) the immediate post-BA gradient; and 8) procedural complications.

**Long-term survival.** In order to investigate the long-term impact of BA as effective treatment for arch obstruction, survival rates for patients who underwent BA for recoarctation were compared with those of patients who underwent a Norwood procedure for HLHS and did not develop re-coarctation of their neo-aorta.

**Statistical analysis.** Continuous variables are expressed as mean ± SD. Comparison of continuous variables was performed using paired Student t tests. Categorical variables were compared using Fisher exact test. Relationships between continuous variables were evaluated with simple as well as multivariant logistical regression models. Freedom
from recurrent coarctation following BA was calculated using Kaplan-Meier curves. Survival differences between those who developed re-coarctation treated by BA and those who never developed re-coarctation were compared by log-rank tests.

RESULTS

Between December 1986 and June 2001, 954 patients with HLHS underwent a Norwood procedure; 633 survived more than 48 h after surgery. In 58 patients (9.2%) re-coarctation of the aorta occurred. The remaining 575 patients served as the control group for this study. Among the re-coarctation population, 23 patients, all before 1988, underwent surgical repair of their neo-aorta and are excluded from this study; the remaining 35 patients underwent trans-catheter BA.

The median age at catheterization was 6.6 months (range 1.9 to 35.6 months). The average peak systolic gradient was 37.2 mm Hg (range 10 to 70 mm Hg). All patients had angiographic evidence of a discrete area of stenosis in the descending aorta, with an average reduction in the cross-sectional diameter of 52% (range 21% to 76%).

Of the 35 patients who underwent balloon angioplasty, 31 patients were diagnosed with re-coarctation after the Norwood operation; the remaining four patients had undergone a cavopulmonary anastomosis before diagnosis. In these four patients, mild arch obstruction was present at catheterization before superior cavo-pulmonary connection (average gradient 10 mm Hg; range 7 to 14 mm Hg). Thirty patients who underwent BA had HLHS (21 of whom had aortic atresia); the remaining 5 patients had unbalanced atrioventricular canal defects (n = 2) or double-outlet right ventricle with mitral atresia or stenosis (n = 3). Weight at the time of the Norwood procedure, the presence of aortic atresia, or the year during which the surgery was performed were not associated with an increased risk for the development of re-coarctation.

Efficacy of BA. Balloon angioplasty was successful in 32 of 35 patients (92%). The approaches for balloon dilation were anterograde in 30 patients (86%) and retrograde in five. BA reduced the gradient to an average of 3.9 mm Hg (range 0 to 21 mm Hg). After BA, the residual stenosis accounted for a total reduction in the cross-sectional diameter of 29% (range 5% to 50%). Complications occurred in seven patients (20%): two had temporary femoral artery occlusion responsive to heparin therapy, two had transient hypotension, two had ventricular dysrhythmias (ventricular tachycardia), and one developed neo-aortic insufficiency and ultimately underwent replacement with a prosthetic valve. There were no BA-related deaths or neurologic complications.

Recurrent obstruction. Recurrent obstruction after balloon dilation occurred in six patients (17%), of whom five underwent redilation. All cases of recurrent obstruction occurred within the first year following the initial BA. The freedom from recurrence was 79% one and five years after angioplasty (Fig. 2). Patients with smaller body surface area at the time of initial BA were more likely to develop recurrent arch obstruction (p < 0.03). No other variables were associated with recurrent obstruction.

Survival analysis. Kaplan-Meier analysis revealed that survival of patients with re-coarctation treated by BA was identical with control patients. The date of surgery correlated with survival in both groups; other variables such as the presence of aortic atresia or birth weight did not. Postoperative survival rates for patients with re-coarctation were 93% at 1 year, 73% at 5 years, and 73% at 7.5 years, compared with the control group survival rates of 92%, 62%, and 62%, respectively (Fig. 3).

DISCUSSION

Re-coarctation of the neo-aorta following a Norwood procedure is common (13–16). In a report of 122 necroscopic heart specimens from patients who died after undergoing a Norwood procedure, re-coarctation was the fourth most common cause of death, occurring in 17 of patients (14%) (17). In our series, re-coarctation occurred in 9.2% of patients, which is consistent with earlier observational studies. We did not find any significant variable that predisposed patients to develop re-coarctation. In a small
subset of patients, the first intervention for recoarctation occurred after superior cavopulmonary connection. All these patients had small gradients at catheterization before second-stage surgery. The increased gradient in some was explained by progressive anatomic obstruction, whereas in the others it may have related more to interim changes in hemodynamic state.

Our success rate for angioplasty was 92%, which is similar to previous reports where rates have ranged from 89% to 100% (2,3,7,18). However, when we investigated the longer term follow-up of this patient group, we found that recurrent obstruction following successful BA occurred in 17% of patients. Aside from a smaller body surface area at the time of the initial BA procedure, we could not find any reliable predictors of recurrent obstruction. We did, however, find that when recurrent obstruction occurred, it happened relatively early and was effectively treated with redilation. In our experience, all cases of recurrent coarctation occurred within the first year following initial BA. The freedom from recurrent obstruction was 79% thereafter, with an average five-year follow-up period.

In our series, the incidence of femoral arterial injury was significantly lower than previously reported rates (4,10,11,18–20). In the majority of patients, BA of the coarctation was performed in an anterograde fashion. This permits the use of relatively large profile catheters without the risk of injury to the femoral artery; in the two instances where femoral artery occlusion was noted, a retrograde approach had been used for dilation. However, it is important to note that dilation of the neo-aorta in an anterograde fashion may carry the risk of injury to the neo-aortic valve. In our series, one patient developed severe neo-aortic regurgitation and ultimately required replacement of the valve with a prosthesis. We believe that complete balloon deflation before removal from the aorta minimizes the risk of this complication.

We were interested in determining whether the presence of re-coarctation affected long-term survival. With a follow-up period up to 10 years, we found that that there was no difference in survival for those treated with BA for re-coarctation compared with patients who did not have neo-aortic arch obstruction.

Interpretation of our results is subject to certain limitations. The study population is a preselected group by virtue of the fact that they all survived the initial months after Norwood in order to undergo a cardiac catheterization. Patients who expired before cardiac catheterization with re-coarctation were not included. In addition, because there were no prospective criteria established for the treatment, or even diagnosis, of re-coarctation, indications for dilation were not uniform and may have changed over the study period. Given the lack of generally accepted criteria for re-coarctation after Norwood, we segregated patients based on treatment for arch obstruction rather than specific anatomic or hemodynamic features. Thus, the control group may include some patients with degrees of arch obstruction similar to patients who underwent treatment. The true incidence of arch obstruction after Norwood remains unknown and will depend on what diagnostic criteria are applied. In addition, success was defined as an after-angioplasty peak-to-peak gradient of ≤10 mm Hg after dilation without reference to systemic blood flow. This may have introduced error in our assessment if cardiac output was significantly lower at the time of the post-angioplasty pressure measurement. Finally, the relatively small sample size in this study limits the power of our analysis.

In summary, our data demonstrate that angioplasty for re-coarctation of the aorta after Norwood is efficacious with low morbidity. Recurrent obstruction may occur within the first year, but it is amenable to repeat dilation. Patients with re-coarctation treated by angioplasty have the same survival as those without re-coarctation. Future work is needed to clarify, first, the impact of, and then the approach to, anatomic arch obstruction with small or absent pressure gradients in patients after Norwood.

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